



NATIONAL GUIDELINE CLEARINGHOUSE™ (NGC™) GUIDELINE SYNTHESIS

INFANT HEARING SCREENING

Guidelines Compared

- American Academy of Pediatrics (AAP). Newborn and infant hearing loss: detection and intervention. Pediatrics 1999 Feb;103(2):527-30 [30 references]
- 2. Joint Committee on Infant Hearing (JCIH). <u>Year 2000 position statement.</u> <u>Principles and guidelines for early hearing detection and intervention programs.</u> Am J Audiol 2000 Jun;9(1):9-29 [165 references]
- 3. United States Preventive Services Task Force (USPSTF). Newborn hearing screening: recommendations and rationale. Am Fam Physician 2001 Dec 15;64(12):1995-9 [20 references]

INTRODUCTION:

A direct comparison of guidelines issued by AAP, USPSTF, and JCIH for infant hearing screening are provided in the tables, below. <u>Table 1</u> compares the scope and the content of each guideline. <u>Table 2</u> compares specific recommendations made by each organization regarding universal newborn infant screening prior to hospital discharge; on-going hearing screening following hospital discharge as a means of surveillance; and screening methods. The rating schemes used to classify the strength of the evidence supporting USPSTF recommendations are provided at the end of <u>Table 2</u>. <u>Table 3</u> compares the potential benefits and harms outlined by the guideline developers associated with the implementation of each of the guidelines.

Following the content comparison in Tables 1-3, the areas of agreement and difference among the guidelines are identified. In general, the sensitivity (proportion of infants with hearing loss identified by the screening test) and the specificity (the proportion of infants without hearing loss that pass the screening test) of different screening methods as well as the cost-effectiveness of universal newborn hearing screening are important factors to consider when evaluating differences among these guidelines. The rationale behind disparate recommendations that cannot be attributed to the evidence base available at the time of guideline development is also explored in the discussion of the areas of disagreement.

Abbreviations used in the text and tables follow:

- ABR, Auditory brainstem response
- AAP, American Academy of Pediatrics
- EOAE, Evoked otoacoustic emissions
- JCIH, Joint Committee on Infant Hearing
- NICU, Neonatal Intensive Care Unit

- OAE, Otoacoustic emissions
- SNHL, Sensorineural hearing lossUNHS, Universal newborn hearing screening
- USPSTF, United States Preventive Services Task Force

	OBJECTIVE AND SCOPE
AAP (1999)	To endorse the implementation of universal newborn hearing To review the primary objectives, important components, ar recommended screening parameters that characterize an effective universal newborn hearing screening program
JCIH (2000)	 To endorse early detection of, and intervention for infants with hearing loss through integrated, interdisciplinary state and national systems of universal newborn hearing screening, evaluation, and family-centered intervention To describe the principles underlying effective early hearing detection and intervention programs To provide guidelines on implementing and maintaining a successful early hearing detection and intervention program
USPSTF (2001)	To summarize the third US Preventive Task Force (USPST recommendations on newborn hearing screening and the supporting evidence, and to update the 1995 recommendations contained in the Guide to Clinical Preventive Services, second edition
	TARGET POPULATION
AAP (1999)	Newborn infants
JCIH (2000)	Infants (birth – 3 years)
USPSTF (2001)	Newborn infants
	INTENDED USERS
AAP (1999)	Physicians, audiologist, speech and language therapists, nurses, nurse practitioners, physician assistants, allied health care practitioners, health plans, other

JCIH (2000)	Physicians, audiologists, speech-language pathologists, nurses, nurse practitioners, physician assistants, allied health care practitioners, health plans, parents, other
USPSTF (2001)	Physicians, nurses, nurse practitioners, physician assistants, speech-language pathologists
	INTERVENTIONS AND PRACTICES CONSIDERED
AAP (1999)	Universal screening for hearing loss among all newborn infants Screening Methods:
	 ABR EOAE Other: Tracking and follow-up elements of a UNHS program Identification and intervention practices On-going evaluation of the universal newborn hearing screening (UNHS) program by state monitored systems
JCIH (2000)	Screening: Universal newborn hearing screening prior to hospital discharge after birth Hearing screening of infants born in alternative birthing facilities (e.g., home births) Ongoing surveillance of high risk infants Screening Methods: OAE (either TEOAE or DPOAE) ABR High-risk indicators Follow-up Evaluation: Audiologic and medical evaluation to confirm hearing loss and identify the type of hearing loss Assessment of language and cognitive abilities Other:
	Treatment with amplification devices (e.g., hearing aids)

	 Medical intervention (e.g., removal of cercum, treatment of otitis media) Surgical intervention (cochlear implants) 		
USPSTF (2001)	Screening: UNHS programs Screening Methods: OAE test ABR test Two-stage testing, such as OAE repeated twice, OAE followed by ABR, or automated ABR repeated twice		
	TABLE 2: SCREENING RECOMMENDATIONS		
	Hearing screening prior to hospital discharge after birth?		
AAP (1999)	The Task Force on Newborn and Infant Hearing endorses the implementation of universal newborn hearing screening. Newborn screening has as its goal that 100% of the target population, consisting of all newborns, will be tested in both ears prior to the age of 3 months, with appropriate intervention prior to the age of 6 months. Screening should be conducted before discharge from the hospital whenever possible.		
JCIH (2000)	 The JCIH recommends universal newborn hearing screening. Universal newborn hearing screening includes the following components: All newborns who receive routine care should have access to hearing screening during their hospital birth admission. All newborns or infants who require neonatal intensive care should receive hearing screening before discharge from the hospital. Newborns in alternative birthing facilities, including home births, should have access to and/or be referred for screening before 1 month of age. For locations where universal newborn hearing screening programs are not yet available or where resources limit the development of a universal newborn hearing screening program, the JCIH has identified risk indicators that can be used to help identify infants (birth to 28 days old) who should receive audiologic evaluations. These indicators are: An illness or condition requiring admission of 48 hours or greater to a NICU. 		

- Stigmata or other findings associated with a syndrome known to include a sensorineural and or conductive hearing loss.
- Family history of permanent childhood sensorineural hearing loss.
- Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
- In utero infection such as cytomegalovirus, herpes, toxoplasmosis, or rubella.
- All infants who do not pass the birth admission screen and any subsequent rescreening should begin appropriate audiologic and medical evaluations to confirm the presence of hearing loss before 3 months of age. The infant should be referred for comprehensive audiologic assessment and specialty medical evaluations to confirm the presence of hearing loss and to determine type, nature, options for treatment, and (whenever possible) etiology of the hearing loss.
- All infants with confirmed permanent hearing loss should receive services before 6 months of age.

USPSTF (2001)

- The USPSTF concludes the evidence is insufficient to recommend for or against routine screening of newborns for hearing loss during the postpartum hospitalization. (*I-Recommendation*)
- The USPSTF found good evidence that newborn hearing screening leads to earlier identification and treatment of infants with hearing loss. However, evidence to determine whether earlier treatment resulting from screening leads to clinically important improvement in speech and language skills at age 3 years or beyond is inconclusive because of the design limitations in existing studies.
- Although earlier identification and intervention may improve
 the quality of life for the infant and family during the first year
 of life, and prevent regret by the family over delayed
 diagnosis of hearing loss, the USPSTF found few data
 addressing these benefits. The USPSTF could not determine
 from existing studies whether these potential benefits
 outweigh the potential harms of false-positive tests that many
 low-risk infants would experience following universal
 screening in both high- and low-risk groups.
- The USPSTF found good evidence that the prevalence of hearing loss in infants in the newborn intensive care unit (NICU) and those with other specific risk factors* is 10 to 20 times higher than the prevalence of hearing loss in the general population of newborns. Both the yield of screening and the proportion of true positive results will be substantially higher when screening is targeted at these high-risk infants, but selective screening programs typically do not identify all infants with risk factors. Evidence that early identification and intervention for hearing loss improves speech, language, or

auditory outcomes in high-risk populations is also limited. *Risk factors for sensorineural hearing loss (SNHL) include NICU admission for 2 days or more; syndromes known to include hearing loss (e.g., Usher's syndrome, Waardenburg's syndrome); family history of childhood SNHL; congenital infections (e.g., toxoplasmosis, bacterial meningitis, syphilis, rubella, cytomegalovirus, herpes virus); and craniofacial abnormalities (especially morphologic abnormalities of the pinna and ear canal). Routine hearing screening following hospital discharge (surveillance) AAP Physicians should provide recommended hearing screening, not only (1999)during early infancy but also through early childhood for those children at risk for hearing loss (e.g., history of trauma, meningitis) and for those demonstrating clinical signs of possible hearing loss. **JCIH** All infants who pass newborn hearing screening but who (2000)have risk indicators for other auditory disorders and/or speech and language delay receive ongoing audiologic and medical surveillance and monitoring for communication development. Infants with indicators associated with lateonset, progressive, or fluctuating hearing loss as well as auditory neural conduction disorders and/or brainstem auditory pathway dysfunction should be monitored. The JCIH recommends the following indicators for use with neonates or infants (29 days through 2 years). These indicators place an infant at risk for progressive or delayedonset sensorineural hearing loss and/or conductive hearing loss. Any infant with these risk indicators for progressive or delayed-onset hearing loss who has passed the birth screen should, nonetheless, receive audiologic monitoring every 6 months until age 3 years. These indicators are as follows: Parental or caregiver concern regarding hearing, speech, language, and or developmental delay. Family history of permanent childhood hearing loss. Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian tube dysfunction. Postnatal infections associated with sensorineural hearing loss including bacterial meningitis. In utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis. Neonatal indicators-specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation.

Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's syndrome. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome. Head trauma. Recurrent or persistent otitis media with effusion for at least 3 months. Because some important indicators, such as family history of hearing loss, may not be determined during the course of UNHS programs, the presence of all late-onset risk indicators should be determined in the medical home during early wellbaby visits. Those infants with significant late-onset risk factors should be carefully monitored for normal communication developmental milestones during routine medical care. **USPSTF** No recommendation offered. (2001)What type of screening test should be used? **AAP** The methodology should detect, at a minimum, all infants (1999)with significant bilateral hearing impairment, i.e., those with hearing loss > 35-decibel in the better ear. The methodology used in screening should have a falsepositive rate, i.e., the proportion of infants without hearing loss who are labeled incorrectly by the screening process as having significant hearing loss, of < 3%. The referral rate for formal audiologic testing after screening should not exceed The methodology used in screening ideally should have a false-negative rate (i.e., the proportion of infants with significant hearing loss missed by the screening program), of zero. Until a specific screening method(s) is proved to be superior, the Academy defers recommendation as to a preferred method. Currently, acceptable methodologies for physiologic screening include EOAE and ABR, either alone or in combination. Both methodologies are noninvasive, quick (< 5 minutes), and easy to perform, although each assesses hearing differently. Although EOAE screening is even quicker and easier to perform than ABR, EOAE may be affected by debris or fluid in the external and middle ear, resulting in referral rates of 5% to 20% when screening is performed during the first 24 hours after birth. ABR screening requires the infant to be in a quiet state, but it is not affected by middle or external ear debris. Referral rates < 3% may be achieved when screening is

performed during the first 24 to 48 hours after birth. Referral rates < 4% are generally achievable with EOAE combined with automated ABR in a two-step screening system or with automated ABR alone. In a two-step system using EOAE as the first step, referral rates of 5% to 20% for repeat screening with ABR or EOAE may be expected. The second screening may be performed before discharge or on an outpatient basis within 1 month of age.

JCIH (2000)

- All infants should have access to hearing screening using a
 physiologic measure. Current physiologic measures used for
 detecting unilateral or bilateral hearing loss of various
 severities include OAEs, either transient-evoked (TEOAE) or
 distortion-product (DPOAE), and/or ABR. Both OAE and ABR
 technologies have been successfully implemented for
 universal newborn hearing screening.
- Screening technologies that incorporate automated response detection are preferred over those that require operator interpretation and decision making.
- Programs that use trained and supervised nonprofessional staff must use technologies that provide automated passrefer criteria.

USPSTF (2001)

- Two types of tests are commonly used: otoacoustic emissions (OAE) and auditory brainstem response (ABR).
 Typically, screening programs use a two-stage approach (either OAE repeated twice, OAE followed by ABR, or automated ABR repeated twice).
- The true sensitivity and specificity of newborn hearing screening are difficult to estimate from most screening programs. One-stage screening with an ABR or OAE test can detect 80 to 95% of affected ears, depending on how an abnormal test result is defined. The two-stage protocol of OAE and ABR missed 11% of affected ears, but was more specific than testing with the ABR of OAE alone. Because the prevalence of sensorineural hearing loss is low, there are many more false positives than true positives, especially in low-risk populations.
- If a program for routine hearing screening of newborns is implemented, it should include systematic education to fully inform parents and clinicians about the potential benefits and harms of the testing protocol. Most infants with positive inhospital screening tests will subsequently be found to have normal hearing, and clinicians should be prepared to provide reassurance and support to parents of infants who need followup audiologic evaluation.
- If any program for newborn hearing screening is implemented, screening should be conducted using a validated protocol, usually requiring two screening tests.
 Equipment used should be well maintained, staff should be thoroughly trained, and quality control programs to reduce

	avoidable false-positive tests should be in place. Programs should develop protocols to ensure that infants with positive screening tests receive appropriate audiologic evaluation and follow-up after discharge.
	Rating Scheme
AAP (1999)	Not applicable
JCIH (2000)	Not applicable
USPSTF (2001)	The U.S. Preventive Services Task Force (USPSTF) grades its recommendations according to one of five classifications (A, B, C, D, or I), reflecting the strength of evidence and magnitude of net benefit (benefits minus harms).
	A The U.S. Preventive Services Task Force (USPSTF) strongly recommends that clinicians routinely provide [the service] to eligible patients. (The USPSTF found good evidence that [the service] improves important health outcomes and concludes that benefits substantially outweigh harms.)
	B The U.S. Preventive Services Task Force (USPSTF) recommends that clinicians routinely provide [the service] to eligible patients. (The USPSTF found at least fair evidence that [the service] improves health outcomes and concludes that benefits outweigh harms.)
	C The U.S. Preventive Services Task Force (USPSTF) makes no recommendation for or against routine provision of [the service]. (The USPSTF found at least fair evidence that [the service] can improve health outcomes but concludes that the balance of benefits and harms it too close to justify a general recommendation.)
	D The U.S. Preventive Services Task Force (USPSTF) recommends against routinely providing [the service] to asymptomatic patients. (The USPSTF found at least fair evidence that [the service] is ineffective or that harms outweigh benefits.)
	I The U.S. Preventive Services Task Force (USPSTF) concludes that the evidence is insufficient to recommend for or against routinely providing [the service]. (Evidence that [the service] is effective is lacking, of poor quality, or conflicting and the balance

of benefits and harms cannot be determined.)

TABLE 3: BENEFITS AND HARMS

POTENTIAL BENEFITS POTENTIAL HARMS AAP False Positives: A Screening by high-risk (1999)registry alone (e.g., proportion of infants family history of without hearing loss will deafness) can only be labeled incorrectly by identify ~50% of the screening process as newborns with significant having significant hearing congenital hearing loss. loss. These infants will Reliance on physician require additional testing. observation and/or The goals of universal parental recognition has screening programs not been successful in include maintaining this the past in detecting false-positive rate at < significant hearing loss in 3% and the referral rate the first year of life. for formal audiologic Universal screening has testing after screening at as its goal that 100% of < 4%. infants with significant congenital hearing loss shall be identified by 3 months of age and shall have appropriate and necessary intervention initiated by 6 months of age. JCIH Early detection and False Positives: The (2000)intervention will serve to OAE is known to be maximize linguistic and sensitive to outer ear communicative canal obstruction and competence and literacy middle ear effusion, and, development for children therefore, temporary who are hard of hearing conductive dysfunction or deaf. Without can cause a positive test result (a "refer" outcome) appropriate opportunities to learn language. in the presence of normal children who are hard of cochlear function. hearing or deaf will fall False Negatives: Some behind their hearing infants with hearing loss will pass the newborn peers in language, cognition, and socialhearing screening. Both emotional development. auditory brainstem Such delays may result response and OAE in lower educational and technology can show employment levels in false-negative findings. In adulthood. addition, because OAE responses are generated

within the cochlea by the outer hair cells, OAE evaluation does not detect neural (i.e., eighth nerve or auditory brainstem pathway) dysfunction. Infants with auditory neuropathy or neural conduction disorders without concomitant sensory (i.e., outer hair cell) dysfunction will not be detected by OAE.

USPSTF (2001)

 Effectiveness of Early Intervention to Improve Language Outcomes

> There are no prospective, controlled studies that directly examine whether newborn hearing screening and earlier intervention result in improved speech, language, or educational development. Although several retrospective studies have variously concluded that infants entering treatment programs at younger ages, or infants identified in hospitals with universal screening programs, have better long-term language outcomes, all of these studies have significant methodological flaws.

 Other Potential Benefits of Screening and Treatment

> Because universal newborn hearing screening reduces the average age for intervention by 6 to 9 months, improved

• False Positives:

Because most positive screening tests are false positives, the most likely potential adverse effects of screening are parental anxiety and misunderstanding, and labeling of normal infants as hearing-impaired until the definitive diagnosis can be made months later. Even a small increased risk of these effects could have a large impact on the net benefit of a screening program. In low-risk populations, there are 25 to 50 false positives for each true case of hearing impairment. In existing newborn hearing screening programs, 13% to 31% do not follow up for definitive testing, which might allay concerns about the baby's health.

Findings from studies that evaluated parental anxiety are mixed. In the largest controlled trial of screening, parents whose infants were screened had similar

hearing or increased prelanguage stimulation over that period might, in themselves, be considered important benefits of newborn hearing screening. In addition, there might be a psychological benefit to parents or to hearingimpaired children of avoiding regret in the future due to the delayed diagnosis and treatment of hearing impairment. However, the USPSTF was unable to identify any evidence that would allow it to assess the magnitude of these potential benefits or determine whether they alone were sufficient to offset the potential harms of screening.

anxiety and attitudes as parents whose infants were not screened. In another survey, 98% of parents said they would give permission for screening, 95% said they would prefer screening even if the baby failed, and 85% said that anxiety caused by failing a screening test would be outweighed by the potential benefit of early detection. In other studies, false-positive results produced significant or lasting anxiety in 3% to 14% of parents, even after follow-up testing. No studies have evaluated whether parental anxiety has any long-term effect on parent-child interaction.

Because definitive diagnoses may take months to confirm, falsepositive diagnosis of sensorineural hearing loss may occasionally lead to unnecessary intervention in an infant who hears normally. In one large screening trial, the initial audiologic diagnosis was incorrect in 2 of 27 infants diagnosed with sensorineural hearing loss (7%), and the infants proved to have normal hearing when reexamined at age 4 months or 10 months.

GUIDELINE CONTENT COMPARISON

A direct comparison of guidelines issued by AAP, USPSTF, and JCIH for infant hearing screening are provided in Tables 1-3 above. All three guidelines focus on recommendations regarding universal newborn hearing

screening (UNHS) prior to hospital discharge and discuss the types of screening tests used. In addition, each of these organizations provide explicit reasoning behind their judgments. JCIH also provides recommendations regarding the role of the parent and/or primary care physician in ongoing surveillance for hearing impairment following hospital discharge.

In addition to infant hearing screening, both AAP and JCIH present recommendations for the tracking and follow-up elements of universal newborn hearing programs. AAP also presents recommendations for the ongoing evaluation of state-monitored systems. Lastly, JCIH presents recommendations for treatment and medical intervention following the diagnosis of a hearing loss.

In the following sections, the areas of agreement and difference between the guidelines are evaluated with respect to: 1) hearing screening in early infancy; 2) the issue of UNHS prior to hospital discharge; 3) ongoing surveillance in the primary care setting and through parental concern; and 4) recommended screening methods.

Areas of Agreement

Hearing screening in early infancy

Guidelines issued by AAP and JCIH are in general agreement with the policy of screening infants for hearing loss either prior to hospital discharge. USPSTF concluded that the evidence was insufficient to recommend for or against routine screening of newborns for hearing loss during the postpartum hospitalization (see Areas of Differences). AAP and JCIH endorse universal newborn hearing screening, with a goal of identifying all infants with hearing loss prior to the age of 3 months and initiating treatment by 6 months. Because of the unique accessibility of almost all infants in the newborn nursery (excepting those born in alternative birthing facilities such as the home), both of these organizations recommend that screening take place prior to hospital discharge after birth. AAP and JCIH also recommends that screening be available for all out-of-hospital births.

Routine hearing screening following hospital discharge (surveillance)

JCIH recommends that all infants who pass newborn hearing screening and who also have specific risk indicators (see recommendations in Table 2 above) receive audiologic monitoring every 6 months until age 3 years. AAP likewise recommends that physicians provide hearing screening throughout early childhood for those infants at increased risk for hearing loss (e.g., history of trauma, meningitis) and for those demonstrating clinical signs of possible hearing loss. USPSTF does not address routine hearing screening of infants following hospital discharge in its guideline, although it does support the development of clinical protocols to ensure that infants with positive screening tests receive appropriate testing and follow-up after discharge.

Areas of Differences

Universal infant hearing screening prior to hospital discharge

Recommendations by USPSTF, AAP, and JCIH differ regarding the endorsement of universal newborn screening prior to hospital discharge. As noted above. AAP and JCIH recommend the implementation of universal screening prior to hospital discharge after birth in all infants based on evidence that undetected hearing impairment during infancy and early childhood interferes with the development of speech and verbal language skills. USPSTF on the other hand, states that there is insufficient evidence to recommend for or against routine screening of newborns for hearing loss during the postpartum hospitalization, noting that evidence is inconclusive as to whether earlier treatment resulting from screening leads to clinically important improvement in speech and language skills at age 3 or beyond. USPSTF does not dispute that reduced hearing during infancy is associated with developmental delay in speech and language. They simply state that no controlled clinical trials have vet evaluated the efficacy of early screening in terms of long-term function and quality of life outcomes. In addition, USPSTF states that current screening methods (ABR and EOAE), while having reasonably high sensitivity and specificity, yield a substantial number of false positive results because of the low prevalence of hearing loss in low-risk infants. JCIH and AAP note that a policy of screening only high-risk infants will neglect approximately 50% of infants with hearing loss.

Screening methods

There are differences between the guidelines with respect to the screening technology that is endorsed. JCIH recommends that all infants have access to screening using a physiologic measure (either otoacoustic emissions [TEOAE or DPOAE] and/or auditory brainstem response [ABR]). AAP states that although additional research is necessary to determine which screening test is ideal, EOAE and/or ABR are presently the screening methods of choice. AAP defers recommending a preferred screening test. USPSTF also does not recommend a specific screening test, but it does cite data on the sensitivity and specificity of OAE and ABR using visual reinforcement audiometry (performed at 8 to 12 months) as the "gold standard." In the cited study, onestage screening with an ABR or OAE test detected 80-95% of affected ears, and a two-stage protocol of OAE and ABR missed 11% of affected ears. The two-stage protocol, however, was more specific than testing with either ABR or OAE alone. The number of false-positives with both protocols, however, was very high, especially in low-risk populations. USPSTF, therefore, recommends that if a newborn hearing screening is implemented, the screening should be conducted using a validated protocol, usually with two screening tests. Well-maintained equipment, thoroughly trained staff, and quality control programs are also recommended to avoid false-positive tests.

This Synthesis was prepared by ECRI on February 15, 2001. It was reviewed by the guideline developers as of May 24, 2001. It was modified on November 20, 2001 to reflect the updated USPSTF recommendations and the removal of two guidelines from the NGC Web site (one from the Office of Medical Applications of Research and the other from the Canadian Task Force for Preventive Health Care).

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